INTRODUCTION

Vestibular schwannoma is a relatively uncommon tumor. Although it is benign but locally expansile and spreads to damage the adjacent structures. Treatment strategy includes surgery, Stereotactic Radiosurgery (SRS) either by standard or hypofractionated protocols. Due to its benign nature, radiation therapy cannot remove the tumor completely, instead radiation therapy halts the growth of vestibular schwannoma and inactivates this benign tumor. Response of radiation in the form of tumor shrinkage is seen 2 - 2.5 years after the radiations. We report a case of vestibular Schwannoma in which residual tumor of 3.1 cm size following subtotal resection was irradiated of the dose of 54 Gy in 30 equal fractions on 3-Dimensional Conformal Radiation Therapy (3-DCRT). A follow-up CT scan brain after 2 months of radiotherapy showed complete disappearance of the disease categorized as complete response. This is an unusual phenomenon and is likely due to the very rarely seen malignant transformation or presence of malignant component in this benign tumor.

Key Words: Vestibular Schwannoma. Radiation therapy. Complete response. 3-D Conformal Radiation Therapy (3-DCRT).

CASE REPORT

Vestibular Schwannoma: An Unusual Post Radiotherapy Response

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ABSTRACT

Vestibular schwannoma is a relatively uncommon tumor. Although it is benign but locally expansile and spreads to damage the adjacent structures. Treatment strategy includes surgery, Stereotactic Radiosurgery (SRS) either by standard or hypofractionated protocols. Due to its benign nature, radiation therapy cannot remove the tumor completely, instead radiation therapy halts the growth of vestibular schwannoma and inactivates this benign tumor. Response of radiation in the form of tumor shrinkage is seen 2 - 2.5 years after the radiations. We report a case of vestibular Schwannoma in which residual tumor of 3.1 cm size following subtotal resection was irradiated of the dose of 54 Gy in 30 equal fractions on 3-Dimensional Conformal Radiation Therapy (3-DCRT). A follow-up CT scan brain after 2 months of radiotherapy showed complete disappearance of the disease categorized as complete response. This is an unusual phenomenon and is likely due to the very rarely seen malignant transformation or presence of malignant component in this benign tumor.

Key Words: Vestibular Schwannoma. Radiation therapy. Complete response. 3-D Conformal Radiation Therapy (3-DCRT).
SRS but due to size (> 3 cm) it was difficult to give SRS treatment. This patient was then simulated in supine position in head mask shell on dedicated CT simulator Siemens 24 slice, volumes contoured on dosimetrist and planned on prowess panther V. 4.2 on 3-Dimensional Conformal Radiation Therapy (3-DCRT) systems in AEMC. Pre-operative MRI scan based tumor volume was recognized and a Gross Tumor Volume (GTV) was drawn. A margin of 2 cm as clinical target volume (CTV) and 0.5 cm Planning Target Volume (PTV) was drawn around GTV with the collaboration of an experienced Radiologist and a radiation Oncologist. Patient was planned by two wedged parallel opposed and one posterior 6 MV photon beams keeping the tumor dose to PTV 54 Gy at 1.8 Gy per fraction shown in Figures 3 and 4 showing position of beams and digital reconstructed radiographs. Surrounding structures were in normal tolerance dose limits as shown in Figure 5 i.e. Mean doses to optic chiasm 30 Gy, brain stem 43 Gy, left optic nerve 35 Gy, right optic nerve 12 Gy, spinal cord 11 Gy and both eyes and lens were in tolerance limits. Voluming and planning were discussed in multi-disciplinary team meeting with other Radiation Oncologists cross opinion. Patient was delivered radiation therapy on linear accelerator ONCOR. Mild dermatitis was observed during the treatment and was treated accordingly. Patient was alright and hearing was improved further.

After 8 weeks of termination of radiation therapy, a CT scan with contrast was done and showed defect in frontal bone with VP shunt in the occipital bone consistent with previous surgery, left mild scleroses of mastoid air cells consistent with postoperative changes/edema changes. Post-contrast images showed no abnormal enhancement. No focal area of abnormal density is seen with symmetrical dilatation of 4th ventricles and was concluded as no residual tumor seen with postoperative changes. On examination, hearing and facial weakness were improved. Patient was followed-up for 3 months and no other abnormal findings were observed. Patient hearing was improved.

**DISCUSSION**

Vestibular schwannoma is a benign tumor. It involves schwann cells in myelin sheath. The treatment of Vestibular schwannoma is surgery and radiation therapy. After radiation therapy, tumor does not disappear but inactivated and manifested as low density mass on CT scan as compared with previous CT scan. In this case, postoperative residual vestibular schwannoma was treated with 3-D Conformal Radiotherapy and delivered 54 Gy dose. For large size tumor where gross total resection may cause nerve damage i.e. facial or glossopharyngeal nerves, subtotal resection may be appropriate. It was found that tumor totally disappeared after radiation therapy. This phenomenon was likely due to the malignant transformation of the benign tumor cells or presence of large malignant component which responded very well but there was no histological confirmation.

Only a handful of cases of de-novo malignancies of the vestibulocochlear nerve have been reported. Even rarer is the malignant transformation of a previously histologically diagnosed benign vestibular schwannoma. There is a reported case of a young adult who developed malignant vestibular schwannoma after combined operative/Gamma knife treatment for a benign vestibular schwannoma. 7

Evaluating the cause of malignant transformation of a benign tumor is made more challenging by the fact that malignant transformation is part of the natural history of a small proportion of benign tumors, including about 0.14% of vestibular schwannomas not associated with...
NF-2\textsuperscript{2} although no studies have definitively shown this instability in human cancers. One case report showed the mutation of the p53 tumor suppressor gene in a malignant transformed vestibular schwannoma.\textsuperscript{9}

The optimal management of malignant vestibular nerve tumors, regardless of origin, is uncertain. Several combinations of aggressive surgical resection and adjuvant radiotherapy and chemotherapy have been attempted with poor clinical outcome. The median life expectancy after diagnosis of malignant vestibular schwannomas or MPNST of the vestibulocochlear nerve is 5 - 8 months, although some patients have survived 24 months. Systemic metastasis has been described.\textsuperscript{10}

Since SRS is not available everywhere in Pakistan and most of the radiation simulation is done on two dimensional fluoroscopy machine (2D). Three dimensional conformal techniques are useful and effective where SRS is not available.

It is concluded that malignant transformation or malignant component of the benign tumor are very rare in vestibular schwannoma and may not be associated with neurofibromatosis or previous radiation exposure and responded well to radiation therapy on three dimensional conformal radiotherapy.

REFERENCES